

New Classification of Headache

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SUMMARY

The Headache Classification Committee of the International Headache Society has developed a new classification system for headache, cranial neuralgia, and facial pain. The value of the classification for the practising clinician is that it forces him or her to take a more careful history in order to determine the nature of the headache. This article reviews the classification system and gives examples of case histories and subsequent diagnoses.

RÉSUMÉ

Le Comité de classification des céphalées de la Société internationale des céphalées a développé un nouveau système de classification des céphalées, névralgies crâniennes et douleurs faciales. Pour le clinicien en pratique, cette classification a pour effet de le forcer à obtenir une histoire plus précise s'il désire déterminer la nature de la céphalée. Cet article passe en revue ce système de classification et l'illustre par des histoires de cas et des diagnostics qui en résultent.

Can Fam Physician 1992;38:2062-2066.

THE VALUE OF CLASSIFICATION was well understood by Lewis Carroll in *The Hunting of the Snark*. A brave band of hunters was searching for an ephemeral creature, which came in a variety of subspecies. The lesser snark could be caught by diverse methods, such as railway shares. There was, however, a form of snark that was more deadly, whose victim would quietly fade away. This, of course, is what happened to one of the heroes, who did not hear the bellman's instructions on how to classify the creatures properly.

The management of headaches is much the same. The clinician is dealing with an ephemeral entity, with seemingly diffuse signs. Yet there are myriad symptoms and even signs, which can be interpreted if the physician listens to the patient and examines carefully in a way that is not usually proposed in most textbooks.

The Headache Classification Committee of the International Headache Society (IHS)¹ has developed a new classification system for headache, cranial neuralgia, and facial pain. Like many classifications, it is not perfect; because of this, the committee welcomes suggestions and holds regular meetings to update the system.

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The classification is reproduced here, courtesy of the International Headache Society. Following publication of the classification, numerous attempts were made to test its ability to differentiate different types of headaches. In general, it fits reality better than would be expected, remembering that "reality" is based on clinical signs and symptoms elicited subjectively. It is important that the patient's history be taken carefully and the examination performed specifically to elicit headache signs. The neurological examination is designed to find lesions in the central nervous system. It was devised at the turn of the century, both in Germany and Britain. Unfortunately, the neurological examination, while allowing exclusions of significant neurological disease, does not allow a full assessment of headache.

Examination of the headache patient

Headache is said by Moskovitz² to be a visceral pain, poorly localized. In my opinion and in that of others, it is both visceral and somatic, and there are specific areas where pain can be elicited.

The patient's history is all-important and should lead to diagnosis in 90%³ of cases. Examining the patient should include a neurological examination but also examination of the part of which the patient complains. The fact that the pain resides in the head should not save this organ from a thorough examination. The

temporal arteries must be palpated, or pressed. Many patients will have tenderness on the side ipsilateral to the headache. The occipital areas, especially occipital nerves, are often tender ipsilateral to the headache. Occipital nerve irritation is extremely common in cases of otherwise typical common migraine. The suborbital and supraorbital areas should be examined, as should the temporomandibular joint. The trapezius muscle is often tender in a variety of headache disorders.

The history and examination should provide the physician with a picture of the patient's headache. It is interesting that attention paid to this aspect of the examination will produce a much greater degree of satisfaction to patients, who often feel short-changed. Few people presenting with an acute abdomen would be happy to be told they are neurologically normal. While the central nervous system may relay and partly cause the pain of headache, the central and peripheral manifestations must be identified to allow diagnosis and classification.

Classification is not only useful for the researcher but also extremely valuable for the practising physician. Pigeonholing the type of headache the patient has enables appropriate treatment to be instituted and is reassuring to patients, who often feel that their pattern of symptoms is quite unique. Because headache is such a common primary care presentation, it is useful for the family physician to have a working knowledge of the various types, especially because "headache" teaching is almost non-existent in medical schools. Also, the current perception is that physicians are not particularly knowledgeable or receptive to headache patients' particular problems.¹

What is the value of a classification if it does not allow more effective treatment and prognosis? In the IHS classification there is always the rider that organic lesions must be excluded. Does this mean that each patient with a headache needs computed tomography or magnetic resonance imaging? If the central nervous system examination is completely normal and the headaches are chronic, then it is safe to assume that there is no intracranial lesion. If the headache is new, is unusual, or is accompanied by neurological signs, then investigation is mandatory.

The classification consists of terms with diagnostic criteria. These diagnostic criteria

are based on previous clinical experience. I will go through the classification by sections and explain the terminology and concepts.

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1. Migraine

1.1 Migraine without aura

1.2 Migraine with aura

1.3 Ophthalmoplegic migraine

1.4 Retinal migraine

1.5 Childhood periodic syndromes that may be precursors to, or associated with, migraine

1.6 Complications of migraine

1.7 Migrainous disorder not fulfilling these criteria

The first part of the classification covers migraine (see above); here, the aura is used to differentiate common migraine from classic migraine. "The headache of migraine without aura," as common migraine is now termed, includes unilateral pain, a pulsating quality, a spectrum of intensity, and aggravation by physical exertion. Nausea or vomiting is not necessary for the diagnosis, but nausea, vomiting, photophobia, or phonophobia must be present. Other neurological conditions can be present, provided migraine attacks do not occur for the first time soon after their onset.

Classic migraine is now termed "migraine with aura," and the aura may consist of a variety of symptoms, such as visual disturbances, unilateral paresthesia, unilateral weakness, aphasia, or speech difficulty. When the aura is prolonged, the term "complicated migraine" is used. Other forms of migraine are self-explanatory and follow the previous classification.

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2. Tension-type headache

2.1 Episodic tension-type headache

2.2 Chronic tension-type headache

2.3 Tension-type headache not fulfilling these criteria

Tension headache is now called "tension-type" headache (see above) and, I think, poses a transition in thinking. It is now accepted that the previous concept of the genesis of tension headache, relating it to muscle tension in the neck, is inaccurate and that probably tension headache is a part of the migraine syndrome. This was shown quite convincingly by Drummond and Lance,⁵ who analyzed symptoms suffered by patients with so-called migraine and tension headache and found that one could not distinguish between the two

Example 1. A 24-year-old woman presented with a history of unilateral headaches preceded by 15 minutes of a right-sided visual deficit surrounded by scintillating edges. The headache is associated with intolerance to light and noise, is accompanied by nausea, and lasts for 6 hours. This is clearly migraine with aura (Section 1.2).

Example 2. A 40-year-old man has a 15-year history of recurrent headaches that are bilateral. They are present in the morning and last all day. At times the pain is at the back of the head and is described as a "vise." Once or twice a week the pain becomes throbbing in character and very severe; at these times he has light intolerance and nausea.

This could be classified as a chronic tension-type headache (Section 2.2) with secondary migrainous elements (Section 1.1). Thus, in situations where the headache seems to be of a mixed type, two diagnoses are used.

entities. Currier, in a recent editorial,⁶ stressed a wish that the committee had had the courage to omit the word "tension" from the tension-type classification. "Tension-type" headache is further subdivided into entities with and without muscular involvement. This diagnosis, however, requires the use of electromyography and ignores the fact that some patients have increased electromyographic activity in the pericranial muscles but no headache.

So far the IHS classification is only slightly different from the previous ones. In my opinion the insistence that other neurological disease be ruled out ignores the belief of many physicians that migraine should not be a diagnosis of exclusion. However, it is true that most patients end up being investigated "to exclude something serious," and I think this is reasonable. It can provide reassurance for patients who are skeptical of the physicians' clinical acumen.

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3. Cluster headache and chronic paroxysmal hemicrania

3.1 Cluster headache

3.2 Chronic paroxysmal hemicrania

3.3 Cluster headache-like disorder not fulfilling these criteria

One of the most dramatically distinct headache entities is cluster headache (see above). The description of retro-orbital pain, coming in bouts, lasting no more than several hours, associated with ptosis, miosis, lacrimation, and nasal stuffiness is unmistakable. As well as episodic and chronic cluster, the category now includes chronic paroxysmal hemicrania, where the attacks are shorter, are more frequent, occur mostly in female patients, and respond to indomethacin.

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4. Miscellaneous headaches unassociated with structural lesions

4.1 Idiopathic stabbing headache

4.2 External compression headache

4.3 Cold stimulus headache

4.4 Benign cough headache

4.5 Benign exertional headache

4.6 Headache associated with sexual activity

Section 4 deals with miscellaneous headaches. These include an idiopathic, stabbing headache with attacks lasting several seconds, like a series of stabs. It has

been called "ice-pick" headache. These types of headaches can be very alarming, yet are really relatively common and, again, respond to indomethacin.

The second section of the miscellaneous category is external compression headache, such as that found in police officers who wear heavy helmets, or other people wearing heavy headgear. Section 3 is cold stimulus headache, which can be divided into that caused by external application of the stimulus, such as cold or wind, and that caused by internal application, such as eating ice cream. Other headaches in this group are benign cough headache, benign exertional headache, and headache associated with sexual activity. The latter is either dull, explosive, or postural. The "exploding head" syndrome⁷ is not confined to coitus and comes under the category of other miscellaneous headaches. A recent series followed 10 elderly women who had woken in the middle of the night with the feeling that their heads were exploding. All turned out to have negative investigation results and a benign course.

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5. Headache associated with head trauma

5.1 Acute posttraumatic headache

5.2 Chronic posttraumatic headache

Headache associated with head trauma (see above) is classified separately, being of either acute or chronic onset and related to the severity of the injury.

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6. Headache associated with vascular disorders

6.1 Acute ischemic cerebrovascular disease

6.2 Intracranial hematoma

6.3 Subarachnoid hemorrhage

6.4 Unruptured vascular malformation

6.5 Arteritis

6.6 Carotid or vertebral artery pain

6.7 Venous thrombosis

6.8 Arterial hypertension

6.9 Headache associated with other vascular disorder

Section 6 covers new headaches that appear in close temporal conjunction with the onset of a vascular disorder. The headaches can be either of the tension or migraine type. In this section a causal relationship is not implied. However, in some of these, eg, unruptured aneurysm or

Example 3. A 31-year-old man presented with retro-orbital pain. The pain occurred on one side and lasted about an hour at a time, recurring several times a day. Typically it woke him from sleep at 2 AM. The eye reddened and teared, the eyelid drooped, and the nose became congested. The attacks came in bouts lasting 6 to 8 weeks, and he was free from them for several months at a time. The pain was such that he wanted to hit his head against a wall rather than lie still. This is a typical cluster headache of the episodic type (Section 3.1).

Example 4. A 50-year-old woman was beset by continuous bouts of retro-orbital pain lasting several minutes at a time. These came with great frequency and were very severe. The eyelid on the side of the headache almost constantly drooped, and the sclera was red. There was lacrimation and rhinorrhea. The attacks were partly relieved by acetylsalicylic acid.

This is an example of chronic paroxysmal hemicrania (Section 3.2). In theory this should respond completely to indomethacin in a dose of 150 mL daily.

moyamoya disease, the increased occurrence of headache is due to the disease. In other conditions, eg, temporal arteritis, carotid artery dissection, or venous thrombus, the relationship is direct and causal.

A point concerning the temporal arteritis headache needs to be made. A recent clinical survey of the types of headaches associated with temporal arteritis suggests that it can mimic any kind of headache. There is no typical headache of temporal arteritis, although such symptoms as jaw claudication or transient visual losses are not uncommon.

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7. Headache associated with avascular intracranial disorder

- 7.1 High cerebrospinal fluid pressure
- 7.2 Low cerebrospinal fluid pressure
- 7.3 Intracranial infection
- 7.4 Intracranial sarcoidosis and other non-infectious inflammatory diseases
- 7.5 Headache related to intrathecal injections
- 7.6 Intracranial neoplasm
- 7.7 Headache associated with other intracranial disorders

Section 7 consists of headaches of increased and decreased CSF pressure and of headache due to infections and inflammations.

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8. Headache associated with substances or their withdrawal

- 8.1 Headache induced by acute substance use or exposure
- 8.2 Headache induced by chronic substance use or exposure
- 8.3 Headache from substance withdrawal (acute use)
- 8.4 Headache from substance withdrawal (chronic use)
- 8.5 Headache associated with substances but with uncertain mechanism

Section 8 concerns headaches associated with substance abuse, including ergotamine and analgesic abuse headache. These are becoming increasingly recognized as a cause of the chronic daily headache syndrome, where the patient has a headache every day, overuses analgesics or ergot, and cannot be helped by the usual prophylactic medication without withdrawal of these agents. The mechanism is still controversial, but the consensus is that these headaches are indeed distinct entities and

generally do not respond to standard prophylactic medication.

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9. Headache associated with noncephalic infection

- 9.1 Viral infection
- 9.2 Bacterial infection
- 9.3 Headache related to other infection

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10. Headache associated with metabolic disorder

- 10.1 Hypoxia
- 10.2 Hypercapnia
- 10.3 Mixed hypoxia and hypercapnia
- 10.4 Hypoglycemia
- 10.5 Dialysis
- 10.6 Headache related to other metabolic abnormality

Generalized infection can cause headaches, as can metabolic disorders, such as hypoxia, hypercapnia, and renal dialysis.

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11. Headache or facial pain associated with disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth, or other facial or cranial structures

- 11.1 Cranial bone
- 11.2 Neck
- 11.3 Eyes
- 11.4 Ears
- 11.5 Nose and sinuses
- 11.6 Teeth, jaws, and related structures
- 11.7 Temporomandibular joint disease

The diagnosis of headache associated with disorders of the cranium, neck, face, eyes, ears and sinuses, teeth, and mouth requires strict adherence to the principle that disease in these structures must be discovered and that the headache disappears when this disease is treated.

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12. Cranial neuralgias, nerve trunk pain, and deafferentation pain

- 12.1 Persistent (in contrast to ticlike) pain of cranial nerve origin
- 12.2 Trigeminal neuralgia

Neuralgias include optic nerve demyelination pain, herpetic pain, post-herpetic neuralgia, the Tolosa-Hunt syndrome, the neck-tongue syndrome (sudden turning of the head causing pain and numbness in the distribution of the lingual nerve and second cervical root), trigeminal neuralgia, glossopharyngeal neuralgia, and atypical facial pain.

Example 5. A 41-year-old woman visited the emergency department with a history of 2 weeks of sudden jabs of pain starting at the back of the head and shooting forward. Each jab lasted a few seconds and would be repeated many times. She was very alarmed, as she had not experienced this pain before.

This is an example of an idiopathic stabbing headache (Section 4.1). Is this a diagnosis or a "cop out"? While the cause is unknown, the presentation is stereotypical, the outcome benign, and the symptoms usually respond to indomethacin.

Example 6. A 30-year-old man presented with a 2-month history of debilitating headaches, which started on the first occasion after he had been standing for 10 minutes. They gradually got worse until he was forced to lie down. After he had lain down for 5 to 10 minutes, the headaches improved. The pain was resistant to all the usual analgesics. He had had a myelogram performed 3 months previously.

These are typical low pressure headaches due to a CSF leak following the myelogram (Section 7.2). The leak can be demonstrated radiologically, but pragmatically with this history, epidural injection of the patient's own blood to seal the puncture performed by an anesthetist, will usually clear up all the symptoms.

Conclusion

This exhaustive list covers almost all the eventualities of pain arising in the head and neck. The value of the classification for the practising clinician is that it forces more careful history taking in order to determine the nature of the headache. It is my experience that patients are relieved when their seemingly bizarre symptoms are clearly described in an accepted classification. The recognition of certain headache types that respond to, for instance, indomethacin, is of obvious clinical importance.

The use of a comprehensive classification will certainly aid international research, but it is the clinician who will find the system most useful. The publication is more than a classification—it is a handbook for headache spotters. So far, several studies have compared inter-observer reliability using this classification, and the results, in general, have been satisfactory.

I strongly urge all physicians who are interested in headache to obtain a copy of the issue of *Cephalgia* devoted to the classification. It includes an exhaustive set of references about all the syndromes. It can be obtained from the Norwegian University Press. The Canadian Headache Society is endeavoring to obtain copies of the abridged classification for distribution. ■

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